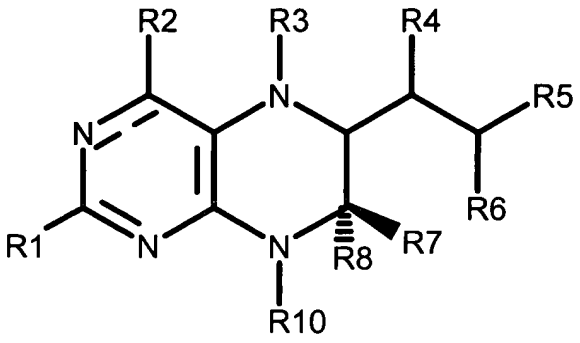
IN THE CLAIMS:

This listing of the claims will replace all prior versions, and listings, of claims in the application:

Claims 1-44 (canceled)

45. (new) A method for long-term treatment of conditions of reduced protein tolerance due to reduced phenylalanine oxidation without deficiency of cofactor tetrahydrobiopterine, said conditions caused by mutations in the phenylalanine hydroxylase gene associated with at least one of the following allele pairs: A403V + IVS4+5G>T, P314S +R408W, F39L + D414N, Y414C + D415N, Y417H + Y417H, F55L + S310Y, V177M + R408W, P275L +Y414C, V245A +R408W, L48S + R158Q, Y417H + Y417H, V245A +R408W, R261X +A300R, R158Q + E390G, Y414C +IVS12+1G>A, I65S +A300S, H170O + A300S, R261Q + Y414C, K274fsdel11bp + E390G, IVS4-5C>G + R480W, I65T + Y414c, E390G + IVS12+1G>A, I65V + R261Q, R158Q + Y414C,

said method comprising administering a medicament containing at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

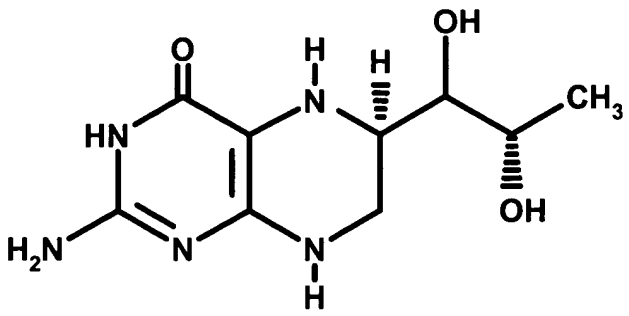
wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond;

as well as their pharmaceutically acceptable salts.

46. (new) A method as in claim 45, wherein said medicament is administered to a patient in need thereof until said patient exhibits improvement in protein tolerance.

47. (new) A method as in claim 45, wherein R1 is NH-acyl, wherein the acyl residue contains CH_3O or 9 to 32 carbon atoms, and wherein at least one of R4 and R6 are C9 to C32 acyl residue.

48. (new) A method according to Claim 45, wherein the compound is selected from the group consisting of: 5,6,7,8- tetrahydrobiopterine, sapropterin, a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone,

and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-etrahydrobiopterine; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.

49. (new) A method according to claim 45, wherein said pharmaceutically acceptable salt is a hydrochloride or a sulphate.

50. (new) A method according to Claim 45, wherein the condition of reduced protein tolerance is at least one of: conditions with elevated phenylaline or reduced tyrosine in body fluids, tissues or cells.

51. (new) A method as in claim 50, wherein said condition of reduced protein tolerance is classic phenylketonurea, mild phenylketonurea, or mild hyperphenylalaninemia.

52. (new) A method according to claim 45, wherein said medicament functions as chaperone for improving protein folding, in particular in the case of structural anomalies of enzymes, which require tetrahydrobiopterine as cofactor.

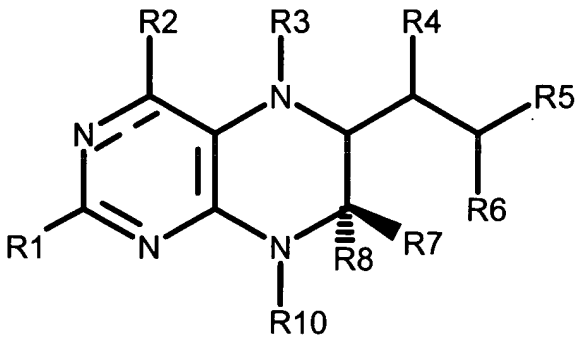
53. (new) A method according to claim 52, wherein said enzyme is selected from phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase and NO-synthase.

54. (new) A method according to claim 45, wherein said compound functions as chaperone as neurotransmitter and/or second messenger enhancer, in particular in conditions with elevated phenylalanine or reduced tyrosin, serotonin, or dopamine in body fluids, tissues or cells, in particular in conditions with reduced phenylalanine hydroxylase, tyrosinhydroxylase, tryptophanhydroxylase or NO-synthase activity.

55. (new) A method according to claim 45, wherein said compound functions as neurotransmitter and/or second messenger enhancer, in particular for catecholamine and/or seratonin and/or dopamine and/or nitrous oxide (NO).

56. (new) A composition containing

(a) at least one compound with the following general formula:



wherein R1 is selected from the group consisting of: H, OH, SH, F, Cl, Br, I, NH_2, $N(CH_3)_2$, $N(C_2H_5)_2$, $N(C_3H_7)_2$; NH-acyl, wherein the acyl residue contains 1 to 32 carbon atoms;

wherein R2 is selected from the group consisting of H, OH, SH, NH_2, F, Cl, Br, I, O, S;

wherein R3 is selected from the group consisting of: H, CH_3, C_2H_5;

wherein R4 and R6 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, acetyl, OX, wherein X is a C1 to C32 acyl residue;

wherein R5 is selected from the group consisting of: phenyl, CH_3, C_2H_5, C_3H_7, butyl, isobutyl, t-butyl;

wherein R7 and R8 are selected independently of each other from the group consisting of: H, OH, SH, NH_2, F, Cl, Br, I, CH_3, COOH, CHO, COOR9, wherein R9 CH_3, C_2H_5, C_3H_7, butyl;

wherein R10 is selected from the group consisting of: H, CH_3, C_2H_5, and -- represents an optional double bond;

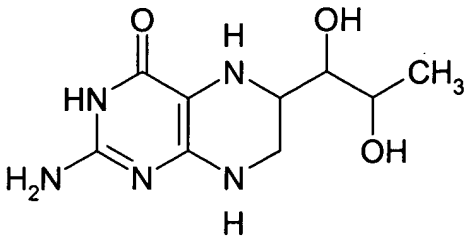
as well as their pharmaceutically acceptable salts, as well as

(b) at least one amino acid selected from the group consisting of

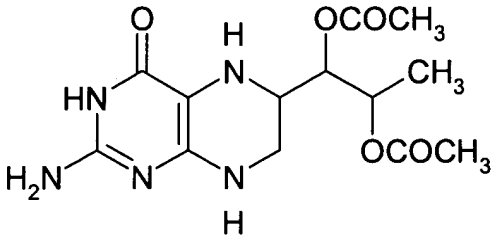
- essential amino acids: isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine; as well as from

- the non-essential amino acids, in particular alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine,

wherein the following compounds are excluded:



and



,

in the case that the amino acid is one of tryptophane, cysteine, in particular acetylcysteine, and tyrosine.

57. (new) A composition according to claim 56, wherein the essential amino acids are selected from the group consisting of isoleucine, leucine, lysine, methionine, threonine, tryptophane, valine, histidine; and that it further contains at least one of the following amino acids: alanine, arginine, asparaginic acid, asparagine, cysteine, in particular acetylcysteine, glutamic acid, glutamine, clycine, proline, serine as well as tyrosine.

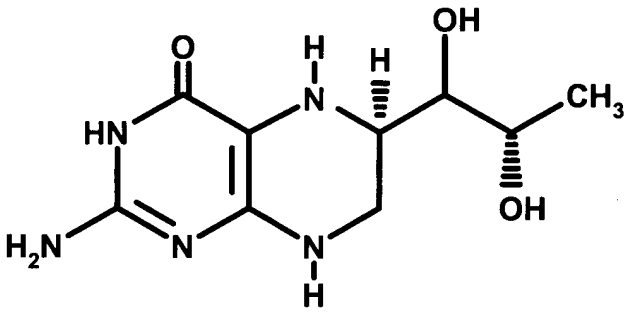
58. (new) A composition according to claim 56, further comprising a hydrocarbon, in particular glucose, and/or vitamins.

59. (new) A composition according to claim 56, formulated as an oral or intravenous preparation.

60. (new) A composition according to claim 59, wherein said composition is in the form of a powder, tablet, capsule, pill, droplets, or as solution for IV administration.

61. (new) A composition according to claim 56, in the form of a pharmaceutical preparation, optionally with pharmaceutical adjuvants or excipients.

62. (new) A composition according to claim 56, wherein the compound is selected from the group consisting of: sapropterin, in particular the hydrochloride thereof, as well as a compound with the following structure:



(-)-(1'R,2'S,6R)-2-amino-6-(1',2'-dihydroxypropyl)-5,6,7,8-tetrahydro-4(3H)-pteridinone, in particular the dihydrochloride or sulphate thereof,

and/or

2-N-stearoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-decanoyl-1',2'-di-O-acetyl-5,6,7,8-etrahydrobiopterine; and/or

2-N-palmitoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine; and/or

2-N-linoleoyl-1',2'-di-O-acetyl-5,6,7,8-tetrahydrobiopterine.